A case of atrial myxoma

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A case of left atrial myxoma in a 9-year-old girl is described. Because of the prominent systemic signs, the diagnosis was only made at necropsy. The importance of early recognition of this condition is stressed.

The myxoma is the commonest primary tumour of the heart in adults, but it is relatively rare in childhood (Van der Hauwaert, 1971). It is usually situated in one or other atrium, more commonly the left. Though about 400 cases have been described, only about 12 atrial myxomata have been reported in children under the age of 12 years (Simcha *et al.*, 1971; Steinke *et al.*, 1972). The tumour was removed in 7 of these, but 2 children died after operation. One further case in a 9-year-old girl is reported here, in whom the condition was not diagnosed until necropsy.

Case report

This girl presented at the age of 9½ years with a history of pain in her right calf over the previous 4 days. Four weeks earlier she had had similar pain in her left calf which had lasted for about 2 weeks. There was a family history of possible rheumatoid arthritis in the mother and maternal grandmother.

On examination, she was mildly obese and afebrile. There was a reddish-violet malar flush, a macular rash at the base of the right hallux, and slight cyanosis of several toes. She had a tachycardia and pronounced sinus arrhythmia, with a soft apical systolic murmur. Her blood pressure was 120/80 mmHg and her peripheral pulses were all present. The muscles of both calves were tender, especially the right. There was no difference in the circumferences of the two limbs.

A provisional diagnosis of dermatomyositis was made from the clinical findings, and the results of some of the investigations were as follows: Hb 13.3 g/100 ml, ESR 65 mm/1 hour Westergren, WBC 6,700/mm³, with a normal differential. Blood urea nitrogen 14.6 mg/100 ml, creatine phosphokinase 77 mU/ml, serum aspartate aminotransferase 40 units Sigma-Frankel (SF), serum alanine aminotransferase 22 units SF; antistreptolysin O titre 833 Todd units/ml. Throat swabs grew no pathogens. Antinuclear factor and Rose-Waaler tests were negative. Chest x-ray showed no abnormality. The electrocardiogram showed sinus arrhythmia, and ST depression in leads V5 and V6 with flattening of the T waves.

She was treated initially with bed-rest and penicillin.

Four weeks after admission she complained of burning pain in the soles of both feet, but apart from a macular rash in this area, nothing could be found to account for the pain. Over the next few weeks her physical signs persisted and corticotrophin gel was given in a dose of 15 units daily initially, increasing to 20 units daily.

Seven months after her initial presentation she developed cardiac failure, which was improved by digoxin therapy. Three months after this she had an episode of pain suggestive of myocardial infarction. Over the next 2 weeks her serum hydroxybutyric dehydrogenase rose to 527 units/ml, and her lactic dehydrogenase to 608 units/ml. Her chest x-ray showed cardiac enlargement and pulmonary congestion and her electrocardiogram (Fig. 1) showed ST segment and T wave changes suggestive of myocardial damage.

During the next few months she had two attacks of acute left hemiparesis with little recovery of function after the second attack. In addition she developed osteoporosis and wedging of her vertebral bodies secondary to the corticotrophin therapy.

FIG. 1 Electrocardiogram shows ST segment and T wave changes.

FIG. 2 Left atrium opened at necropsy showing tumour.

In her final admission she developed pain in her left eye, followed by coma and status epilepticus. Fourteen hours later she died in ventricular fibrillation.

At necropsy the principal finding was a large tumour in the left atrium composed of branching gelatinous fronds, attached by a short stalk to the posterior margin of the foramen ovale (Fig. 2). The tumour consisted of loose connective tissue containing polyhedral and spindle-shaped cells, many of which were multinucleated. There were numerous haemorrhages and deposits of haemosiderin. Staining with Alcian Blue showed small amounts of mucin in the connective tissue. There was no myocardial invasion at the base of the tumour. In the myocardium of the anterior wall of the left ventricle there

was some fibrosis, and a blood vessel was occluded by tumour. Tumour emboli were also found in both middle cerebral arteries and in a small artery in the spleen. The brain showed areas of infarction in the right internal capsule and in the cortex of the right temporal lobe and the right parietal lobe.

Comment

In retrospect the history and clinical findings were suggestive of multiple emboli. The diagnosis of atrial myxoma is often difficult, however, because of the striking systemic symptoms and signs. These have led to an initial clinical diagnosis of rheumatic fever in 7 of the previously reported cases. Of these, only 1 case survived after operation (Steinke *et al.*, 1972), compared with 4 survivors out of the 5 cases where an intracardiac tumour was suspected at an early stage in the illness. Though atrial myxoma is a rare condition in childhood, it is clearly important to consider this diagnosis in cases presenting as rheumatic fever, but showing atypical features.

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References

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